It should also be borne in mind from the point of view of the diagnosis of meningitis that the latter disease may exist especially in alcoholic patients without symptoms. Hoppe-Seiler[21] says that it may occur in old men without stiffness of the neck, vomiting, or motor irritation. In drinkers symptoms may be ascribed to delirium tremens, which are really due to a purulent pneumococcic meningitis of the convexity.

In proportion as one can be sure in his exclusion of the organic group of hemiplegias the prognosis is relatively favorable. We still have the pneumonia to reckon with. But if it is true that the cerebral accident does not materially add to the gravity of the pneumonia, one is justified, on the whole, in giving a rather favorable prognosis to such cases, occurring, as they generally do in young and strong subjects.

SPLENECTOMY FOR SPLENOMEGALY (GAUCHER TYPE).

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This rare type of splenic affection of undetermined etiology received its name following the publication of the doctorate thesis of P. C. E. Gaucher in Paris, 1882, under the title "De l'Epitelioma Primitif de la Rate; Hypertrophie Idiopathique de la Rate Sans Leucemie."[1] Following this, in 1895, Collier[2] published the second case, and in 1896, Picou and Ramond[3] published a case previously reported by the former the year preceding. Bovaird[4] published the initial American case in 1900, this being the fourth on record. The first mention of the condition in German literature was made by Schlagenhaufer in 1907.[5] To date there are sixteen cases recorded in which the clinical diagnosis has been confirmed by operation or autopsy, that of Truelson[6] not being included in the list, because the operative or histological findings do not seem determinative and because no autopsy was performed after the so-called "recurrence."

The earliest recorded splenctomy for this condition (1895) is the case of Picou and Ramond[3] in a patient aged thirty-two years, operated upon for suspected uterine fibroid.

The second splenectomy for authenticated splenomegaly (Gaucher) was a girl, aged thirteen years, operated upon May 17, 1899, by the late A. J. McCosh at the request of Bovaird.[4]

The third case was operated upon in 1905 and is mentioned in an article by von Herczel[7] in 1907, the true nature of the condition not being determined until that time.

Wilson[12] reports the fourth and fifth cases (1907 and 1908) from the Mayo clinic, these having been previously reported as examples of chronic hyperplasia,[33] as stated by Downes in his recent article.[14]

De Jong and Van Heukelom[8] report the sixth case, 1910. In this case the liver continued to enlarge after the splenectomy, and the authors ascribe this to the proliferation of the cells carried there previously from the spleen.

The seventh case (1911) is reported from the Mayo clinic, by Wilson,[12] and this is the first successful American case with an initial pathological diagnosis.

The eighth case was first reported by Reuben[9] in January, 1912, and the operative and autopsy findings (March, 1912) are recorded by Mandelbaum.[10]

The ninth case is reported by Downes,[14] April, 1913.

The tenth case is herewith reported, and in point of age it is the youngest on record. The other case mentioned by us as occuring in the same family was doubtless of the same type, but it is unincluded in the lists because there was no operative or pathological verification.

It is significant that of the ten recorded operative cases there is a family history of the condition in five: Bovaird's, Von Herczel's, De Jong and Van Heukelom's, Reuben and Mandelbaum's, Erdmann and Moorhead's.

Weichselbaum[11] in 1881 reports a splenectomy in a twenty-one year-old soldier; no history is given, but the statement is made that the spleen was enlarged and the title of the article would indicate that the organ was of the Gaucher type. This case is not fully authenticated, and therefore is not recorded with the foregoing.

A tabulation of the recorded cases in the literature has been made by Reuben;[9] and Mandelbaum's[11] and Wilson's[12] recent articles contain a valuable pathological digest of the cases regarded by them as authentic.

The literature now contains some sixteen cases verified by operative or autopsy findings.

The following is a list of the cases submitted to operation:

Priority of publication.	Author and date of publication.	Sex and age.	Blood picture.	Operator and date of operation.	Weight and size.	Outcome.	Duration of enlargement.	Other cases in family.	Priority of operation.
1	Picou and Ramond 1896	Female 32 yrs.	Simple anemia	? 1895	2800 gm.	R.	?	?	1
2	Bovaird 1900	Female 16 yrs.	Simple anemia	McCosh May 17	12½ lbs.	Died in 3 hrs	13 yrs.	Yes	2
3	v. Herczel 1907	Female 37 yrs.	Simple anemia	v. Herczel Sept. 27 1905	3 kg. 32x16 x12 cm.	R.	4 yrs.	Yes	3
4	de Jong and von Heukelom 1910	Female 12½ yrs.	Simple anemia	von Rossem March 29 1909	1850 gm. 26x15 x7 cm.	R.	6½ yrs.	Yes	6
5	Mandelbaum 1912	Male 4½ yrs.	Simple anemia	Wiener March 1 1912	490 gm. 18x9.5 x5 cm.	Died next day	1 yr.	Yes	8
6	Wilson* 1913	Female 27 yrs.	Simple anemia	W. J. Mayo Jan. 7 1907	5280 gm.	R.	?	?	4
7	Wilson* 1913	Female 37 yrs.	Simple anemia	W. J. Mayo May 11 1908	895 gm.	R.	?	?	5
8	Wilson 1913	Female 25 yrs.	Simple anemia	W. J. Mayo Sept. 26 1911	4500 gm.	R.	?	?	7
9	Downes 1913	Female 28 yrs.	Simple anemia	Downes Dec. 14 1912	1813 gm. 35x13.5 x6.5 cm.	R.	15 yrs.	No	10
10	Erdmann and Moorhead 1913	Female 3¼ yrs.	Simple anemia	Erdmann June 6 1912	430 gm. 17x10.5 x7 cm.	R.	2 yrs.	Yes	9

* These cases are not as yet fully authenticated.

SUMMARY.

Female, 9; male, 1.	Recovered, 8; died, 2.
Oldest, 37; youngest, 3.	Other cases in family, 5; unstated, 4; none, 1.
Longest duration, fifteen years; shortest, one year.	

The clinical aspects of the affection have been well described by numerous authors, notably Bovaird,[4] Brill, Mandelbaum, Libmann,[11] Reuben,[9] and best and most recently from a critical pathological standpoint by Mandelbaum.[10] For this reason, and because the diagnosis is relatively plain, we purposely refrain from discussing the medical or clinical features, and revert to the propriety of splenectomy as the treatment of choice, in effect regarding the condition as an operable tumor of the spleen. We are led to advocate operation for the following reasons:

1. All known forms of therapy have hitherto failed, including *x*-ray treatment.

2. Even in advanced cases splenectomy has resulted in relief of symptoms (weight, pressure, gastric and intestinal distress, malnutrition, etc).

3. Removal of the organ is regarded as the method of choice in Banti's disease and splenic anemia, many splenectomies for these and allied conditions now being recorded; hence the analogy seems warranted.

4. The hemogenetic (or ferrogenetic) function of the spleen is apparently unaffected after splenectomy, as indicated by a prompt return of the blood to normal.

5. Early recognition of suitable cases should result in a low operative mortality, as the operation *per se* in this class is not much more hazardous than for other intra-abdominal neoplasms.

Carsten[15] reports a mortality of 18.5 per cent. in a collection of 739 cases of splenectomy for various lesions.

Johnson[16] reports a mortality of 27.4 per cent. in 708 collected cases. These statistics indicate an average operative mortality of about 23 per cent., which seems abnormally high.

W. J. Mayo[13] reports 27 splenectomies (3 of Gaucher ? type), with 2 operative deaths. Of this number 12 remained well one to seven years; 6 were well less than two and a half years. He is of the opinion that the mortality should not exceed 10 per cent.

Wilson,[12] in reporting the pathology of the splenic material from the Mayo clinic, states: "It seems to me unfortunate that the trend of medical thought was originally directed toward the consideration of these endothelial proliferations as true neoplasms, with the associated dread of malignancy, since it has no doubt operated to prevent surgeons from hazarding splenectomy; of the 9 cases which have been operated on, as reported in the literature, 4 have recovered, while the remaining 5 were in a bad condition before coming to operation." Inferentially he intimates that operative interference is as much indicated for splenic connective-tissue overgrowth as for similar pathological changes in the thyroid and prostate.

"Further, the survey of all the (Gaucher) cases suggests a close analogy in their histology with what one meets with in hyperplasias of the thyroid and of the prostate, namely, an initial hyperplasia of the parenchyma filling the acini, a secondary increase of the stroma, and ultimately a degeneration of the parenchyma, with contraction of the connective-tissue."

It appears that ligation of the bloodvessels for various forms of splenomegaly has been practised, but that the mortality is high is indicated by the remark of D. A. K. Steele, who stated that of six cases thus operated upon, four died (discussion of Harris and Herzog's paper).[24]

Cases of enlarged spleen showing a practically normal blood-picture; with a hemoglobin percentage of fifty or more, offer the best prospects for operative interference. Associated pigmentation or marked enlargement of the liver or glands are not contra-indications if the general condition of the patient is good.

Operative complications are generally in the nature of adhesions (intestinal, gastric, pancreatic, parietal, diaphragmatic), adventitious or friable bloodvessels, and attendant shock from manipulation or hemorrhage.

Operative shock and hemorrhage are guarded against by a reasonably large incision and a minimum of intra-abdominal trauma especially to the diaphragm. In motile spleens, primary clamping of the main bloodvessels should be the first step; in other cases, division of the adhesions between clamps is the initial procedure, unless immediate ligation can be more speedily performed. When the organ is large, the bodily blood-supply depleted, or hemorrhage is anticipated, it may be expedient to first expose a vein at the bend of the elbow so that a saline intravenous infusion may be given if necessary. Normally the weight of the spleen in a twelve year-old child is $\frac{1}{400}$ of the body weight; obviously, in a much enlarged spleen, the removal of the organ and the contained blood will cause much depletion, and this can best be counteracted by saline intravenous infusion. Postoperative fever occasionally occurs independent of infection, but ordinarily no special treatment for this is required. It is often easier to deliver the organ from below than from above, and separate ligation of the individual vessels is advisable when practicable.

In the two cases herewith reported it may be stated that the parents are robust and of a highly developed mental and physical type; American born; no constitutional dyscrasiæ.

A genealogical search extending over five generations shows that most of the forebears lived sixty years or more.

The oldest child, Henry, was born March 21, 1901; said to have been an eleven months' pregnancy. Weight, 7 pounds at birth; breast-fed until tenth month. Weight at five months, 13 pounds 10 ounces. Weight at one year, 18 pounds 4 ounces.

Abdominal protrusion first noted about July 2, 1902, and at that time spleen and liver were found enlarged by the family physician, Dr. W. C. Walser, of Staten Island. Blood examination then disclosed malarial parasites, and under appropriate treatment the child seemed to improve.

January 15, 1903. Blood-count (by Dr. H. W. Patterson) showed:

Hemoglobin	62 per cent.
Red blood cells	3,912,000
White blood cells	4,600
Small mononuclears	12.5 per cent.
Large mononuclears	20.0 "
Polynuclears	50.0 "
Basophiles	6.0 "
Eosinophiles	12.5 "

January 22, 1903. Seen by a consultant, who had blood-examination made, which showed a picture similar to the foregoing. A chronic colitis now existed in addition to the splenic and hepatic enlargement.

October 20, 1904. Seen by two other consultants. Various remedies were being constantly administered.

September 18, 1906. Blood-examination by Dr. James Ewing, at request of another consultant, showed practically no changes from the preceding, and a tentative diagnosis of splenic anemia or simple splenomegaly was made.

November 20, 1906. Seen by a former consultant, who stated that the liver and spleen had decreased since the examination of October, 1904.

November 26, 1906. Seen by another consultant, who diagnosticated splenomegaly.

January 1, 1907. Developed pneumonia, with low temperature and much prostration, the cough lasting all spring.

October 27, 1907. Died of pneumonia after one day's illness. No autopsy.

At no time were there any hemorrhages, except slight epistaxis. On one occasion in the latter months "he vomited a great deal, and often looked dark and blackened."

The second child, Frederick, was born February 7, 1903, and is exceptionally strong and well. At the present time he shows no evidences of enlarged spleen or liver.

The third child, William, was born May 8, 1907, and is in splendid health now; no splenic or hepatic involvement.

The fourth child, Elizabeth, is our patient. She was born February 19, 1909; normal delivery; weight at birth, 7½ pounds. Breastfed until seventh month, and then weighed 14 pounds 10 ounces.

November 27, 1909. Had bronchitis; sick for ten days. Weighed 18 pounds at end of first year.

April 8, 1910. Spleen was found slightly enlarged, and at this time a blood-examination was made at the request of Dr. W. C. Walser, of Staten Island, by Dr. H. W. Patterson, and it showed:

Hemoglobin	70	per cent.
Red blood cells	2,956,000	
White blood cells	14,000	
Small mononuclears	13.0	per cent.
Large mononuclears	28.6	"
Polynuclears	41.0	"
Eosinophiles	2.0	"
Basophiles	2.0	"
Myelocytes	12.3	"
Transitionals	1.0	"

At this time she seemed "rather nervous and hard to nourish, the only illness was slight colds."

February 1, 1911. Second blood-count at Dr. Walser's request by Dr. Patterson:

Hemoglobin	83 per cent.
Red blood cells	6,104,000
White blood cells	10,800
Small mononuclears	30.0 per cent.
Large mononuclears	19.0 "
Polynuclears	47.0 "
Eosinophiles	1.0 "
Basophiles	1.0 "
Transitionals	2.0 "

May 27, 1911. Feet badly burned in hot sand, and she was quite ill for several weeks. At this time Dr. Walser noted a considerable increase in the size of the spleen; liver normal.

In July, 1911, she was examined by Dr. R. H. Van Denburg, of Coxsackie, New York, and the splenic enlargement was still present, but the liver was normal. He again saw her September 23, 1911, and found the liver enlarged.

January 24, 1912. Blood-examination showed no special changes from the preceding.

February 21, 1912. Seen by a consultant, and the blood-examination then disclosed:

Hemoglobin (Sahli)	56 per cent.
Red blood cells	5,000,000
White blood cells	9800
Polynuclears	45.0 per cent.
Lymphocytes	55.0 per cent.

Wassermann negative.

At that time the spleen reached to the umbilicus; the liver was smooth, thin edged, and moderately enlarged; no ascites; abdominal veins were enlarged; inguinal glands were enlarged, all others were normal. Splenectomy was suggested if the surgeon justified the risk involved.

February 28, 1912. Blood-examination by Dr. F. E. Sondern at the request of the writers:

Hemoglobin	85 per cent.
Red blood cells (small, varied, and pale)	6,200,000
White blood cells	7000
Small lymphocytes	49.4 per cent.
Large lymphocytes	9.4 "
Polynuclears	40.0 "
Eosinophiles	1.0 "
Basophiles	0.2 "

No malarial plasmodia.

Wassermann negative.

March 2, 1912. Examination by the writers showed a rather pale, poorly developed, somewhat dyspneic child of three years of age. The abdomen was uniformly protuberant and the overlying veins were prominent. No ascites. Liver was smooth, firm, and non-tender, reaching about two inches beneath the costal margin and extending well to the left. Spleen was firm, smooth, non-tender, and reached just below the umbilicus and well toward the median line (Fig. 1). There was one enlarged lymph node in the right inguinal region. No jaundice nor pigmentation. Sclera clear. Bones normal.

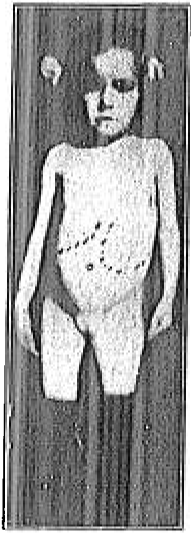
FIG. 1.—Original level of liver and spleen.

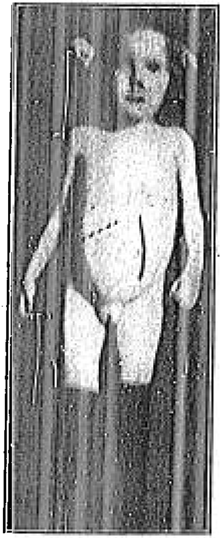
FIG. 2.—Present level of liver and operative scar.

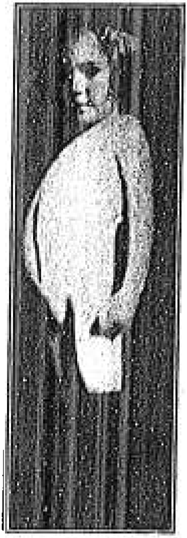
FIG. 3. — Abdominal protrusion and operative scar.

Splenectomy was advised in view of the fatal outcome of the other case in the same family, the apparent progressive loss of strength, and because no improvement had occured despite varied sustained medication. The operation was postponed because of a severe attack of bronchitis accompanied by mastoiditis. Weight just before operation, 26¼ pounds.

June 6, 1912. Splenectomy by Dr. John F. Erdmann assisted by Drs. Moorhead and Russell. Gas-ether anesthesia by Dr. Bennett.

The abdomen was entered through a long incision at the outer border of the left rectus muscle. The spleen was found relatively free, the main attachment being to the splenic flexure and pancreas, and in freeing it from the latter a small portion of the tail of the pancreas was purposely exsected. The splenic vessels were secured by clamps and the organ removed. Separate iodine catgut ligatures for the vessels. Tier sutures of iodine catgut for peritoneum, muscle, and rectus sheath. Continuous lock-stitch suture of the skin. Time of operation, twenty-seven minutes. There was never any temperature over 100°. The patient made a perfect recovery, and left the hospital in two weeks, then weighing 24¼ pounds.

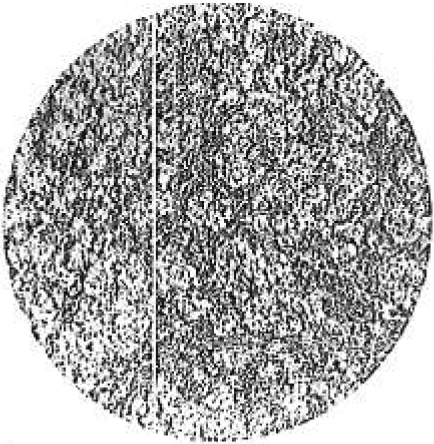

FIG. 4.—Section of spleen showing alveoli filled with large cells. Dr. Sondern.

Following is the histological examination as reported by Dr. F. E Sondern:

The striking lesion microscopically in the spleen is the presence of enormous numbers of large mononuclear cells. These cells are arranged in alveoli throughout the organ, outlined in an irregular manner by a relatively scant connective-tissue network. The nuclei of the cells, which readily stain, are usually centrally situated, but often eccentrically, and occasionally at the border of the cell. The protoplasm is granular and stains faintly. A few of the cells contain vacuoles. Very rarely a cell with two or three nuclei is seen. Connective-tissue fibrils are not apparent between the cells except at the edges of the alveoli, where the cells lie adjacent to the connective tissue.

The endothelium of the venous sinuses is swollen, and occasionally large cells are seen within the lumina, apparently of the same type as those seen elsewhere throughout the pulp.

The Malpighian bodies are few and not increased in size. Scattered among the lymphoid cells of the follicles are a few of appar-

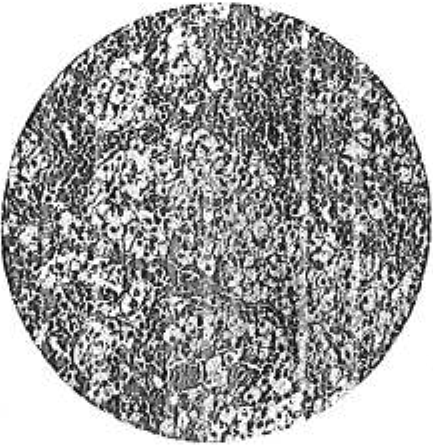

FIG. 5.—Section of spleen showing alveoli filled with large cells. Dr. Sondern. × 500.

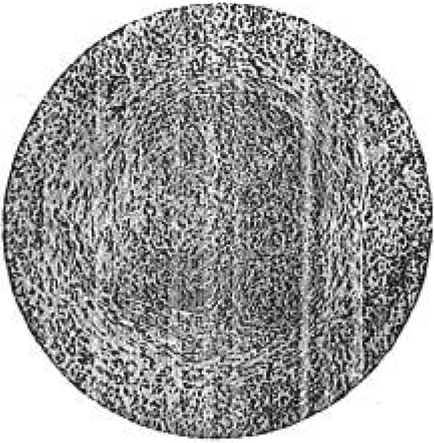

FIG. 6.—Tubercles in spleen. Dr. Mandelbaum

ently the same large mononuclear cells. There are no areas of degeneration or necrosis.

Sections were also made and studied by Dr. F. S. Mandelbaum, who found typical tubercles in addition.

The lesion is a primary splenomegaly of the type first described by Gaucher. It is difficult to determine absolutely the nature of the cells making up the bulk of the tissue. From the morphological appearance of the cells and the swollen condition of the endothelium of the small vessels they are believed to be of endothelial origin.

Spreads made from the surface of the spleen show rather small and pale red cells. There are relatively few leukocytes, with differential count as follows:

Small lymphocytes	74.2	per cent.
Large lymphocytes	7.2	"
Polynuclears	15.6	"
Eosinophiles	3.0	"
Basophiles	none	

June 15, 1912 (nine days postoperative). Blood examination by Dr. Sondern shows:

Hemoglobin	85	per cent.
Red blood cells	6,080,000	
White blood cells	19,000	
Small lymphocytes	24.4	per cent.
Large lymphocytes	20.4	"
Polynuclears	52.6	"
Eosinophiles	2.2	"
Basophiles	0.4	"

July 8, 1912 (one month postoperative). Blood-examination by Dr. Sondern:

Hemoglobin	78	per cent.
Red blood cells	5,200,000	
White blood cells	12,000	
Small lymphocytes	32.4	per cent.
Large lymphocytes	12.0	"
Polynuclears	53.2	"
Eosinophiles	2.0	"
Basophiles	4.0	"

During the summer the child was reported as being much stronger, and far more active than ever before.

September 28, 1912. Examination by one of us showed her to be apparently in good health, the improvement in color being quite noticeable. The protrusion of the abdomen is less marked; no dyspnea; no dilatation of the surface veins; scar firm. Periumbilical circumference, seventeen and three-fourths inches. Liver reaches about one inch below costal border. No inguinal adenitis.

Weight, 26 pounds. Appetite reported as being enormous (Figs. 2 and 3).

March 2, 1913. Reëxamination shows the general appearance to be good. Liver apparently slightly larger than at the previous examination. Peri-umbilical circumference, twenty inches. She is active, strong, and apparently in perfect health.

April 22, 1913 (ten and a half months postoperative). Blood-examination by Dr. Sondern:

Hemoglobin	75 per cent.
Red blood cells	5,300,000
White blood cells	16,000
Small lymphocytes	47.2 per cent.
Large lymphocytes	8.2 "
Polynuclears	43.0 "
Eosinophiles	1.2 "
Basophiles	0.4 "

Note.—October 6, 1913 (sixteen months post operative) the child is reported as well and strong.

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